

A CASE OF HUNTINGTON'S CHOREA WITH REMARKS UPON THE PROPRIETY OF NAMING THE DISEASE "DEMENTIA CHOREICA."¹

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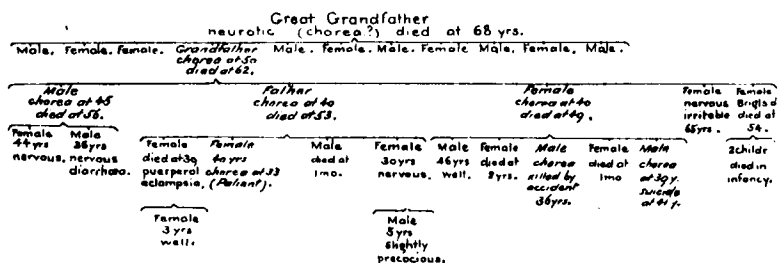
The histories of the majority of reported cases of Huntington's chorea begin rather abruptly with an account of the appearance of the choreiform movements and do not furnish data for the study of the earlier stages of the disease. The following case is, therefore, of interest chiefly because it shows clearly the course and order of development of the degenerative process, the dementia and chorea paralleling each other as the degeneration progresses. The case illustrates a well-recognized type of chronic progressive hereditary chorea, and viewing it from the psychological standpoint, one is strongly inclined to consider the chorea as of secondary importance to the underlying dementia, thereby warranting the descriptive and pathologically more correct name of "dementia choreica."

The patient is the wife of a college professor, native of Massachusetts, 40 years old, and has been under direct personal observation the past year. The accompanying table shows the hereditary history of the patient. It is not known whether the great-grandfather of the patient or his antecedents had chorea. The neuropathic tendencies, however, were transmitted through him. His wife was a very vigorous woman, and of their eleven children only one, it is believed, had the disease. This one, the fourth child, was the grandfather of the patient. He and his two sons used alcohol to excess during their lives. The mother of the patient came of better stock, but was probably never a very strong woman, and after the birth of the patient became a nervous invalid. Beyond what is stated in the diagram of descent, nothing is known of the rest of the family, except that a male cousin of the father was insane and confined

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in the asylum at Taunton, Mass. A glance at the family history shows the strong tendency to degeneration almost to the point of extermination. The reduced number of offspring, early deaths, and the earlier onset and somewhat shorter course of the chorea are significant facts.

The patient was never very strong, but up to the beginning of the disease she enjoyed fairly good health. No rheumatism, tuberculosis, or other special disease is reported in the family. She was married at twenty-one, and has had no children or miscarriages. In 1890, the thirty-second year of her age, the first symptom of the disease showed itself. This consisted of a mental change rather than any choreiform movement. There was, however, a single, isolated motion observed at this time, namely, a rythmical movement of the thumbs during church service. The mental change was the prominent feature and was manifested by a mild depression and lack of confidence in



Note. The names of those affected are in *Italics*.

herself. She did not complain, but seemed to be nervous and fearful of being left alone. If she went shopping without company she would become bewildered and lose her way home. In 1891 and '92 the same lack of confidence continued, and in addition the mental confusion and bewilderment increased. There was more difficulty in comprehension, and she would not talk with people. The motor symptoms accompanying this deepening of the mental condition consisted of no true choreiform movements, but of so-called "choking spells." While seated at the table she would suddenly throw her head back, gasp for breath, and then hurriedly reach for water to drink. Later on these throat spasms occurred in the night and seemed to be semi-hysterical in character.

In 1893 the mental condition of the patient changed. Instead of being afraid to leave home and reluctant to meet people, she now became full of confidence, wanted to travel and visit friends. Before, she was very subdued and anxious

to keep in the background; now she began to feel mildly exalted and pushed herself forward, emulating the acts of others. The "choking spells" at table and the thumb movements in church continued to be noticed with varying frequency. In 1894 the evidence of mental weakness became more noticeable. In ordinary sewing she could not learn to do the simplest things. Her memory began to fail.

In 1895 the first decided choreiform movements appeared. The "choking spells" grew less and were replaced by staggering movements which were first noticed in entering and leaving street cars or carriages. Difficulty in holding objects was experienced, knife, fork and other articles being repeatedly dropped. The menstruation at this time became irregular and remained so a year and a half. Greater mental confusion and failure of memory were observed. In 1896 the movements continued to increase in the extent of muscular involvement. When at rest, e. g., in church, she was quiet, with the exception of occasional arm or leg movements and a rhythmic motion of the head which consisted of looking up at the clergyman and then down to the pew. In walking the irregularity of movement became so great that she appeared to be intoxicated. Disturbance of speech and irregularity in handwriting were noticed. In 1897, her thirty-ninth year, she first came under observation.

Examination.—The patient is a medium sized woman, and, with the exception of the chorea and a rather blank facial expression, presents quite a normal appearance. There are no physical stigmata of degeneration. The heart, lungs and kidneys are normal and her general condition is excellent. There is no evidence of atheroma. The muscular development is uniformly well marked. The menstrual flow is regular, but for the last six months has been scanty. Vision, pupils and fundus normal. Hearing, smell, taste and general sensibility correct. Muscular and temperature senses perfect. No tremor or true ataxia. Romberg symptom absent. Skin reflexes present. Tendon reflexes active, especially knee-jerk, which is slightly exaggerated. No ankle clonus. Electrical reactions normal.

Choreiform Movements.—All muscles of the body are involved, including eyeball, tongue, pharynx, larynx and diaphragm. Patient cannot hold the eyes steady in the presence of another. They shift and wander almost constantly, due in part to irregularity of movements of ocular muscles and in part to motions of head. The tongue is protruded straight and has a large muscular development. It is moved about in the mouth and between the lips very unevenly, causing the char-

acteristic disturbance in articulation, which is labored, hesitating, broken in word sounds and sentences, ending often in a mild explosive utterance. The irregularity of action of the laryngeal muscles and imperfect control of the air column augment the difficulty in articulation. Coughing, choking, and a variety of minor vocal sounds are of frequent occurrence. Swallowing is similarly uneven in its execution. The movements of the facial muscles cause every form of grimace. The trunk and extremities are subject to all possible movements, producing an undulating distortion of the body and limbs from their normal position. The gait is wobbling and uneven, each step varying in its excursion as to height and latitude.

The two sides of the body are equally affected and all movements are greatly intensified in the presence of another person. When lying down or moving about alone the chorea is very much less. All motion ceases during sleep. Volition has a brief effect in quieting the movements. Occasionally the patient is subject (possibly as the result of dreams, certainly from some centrally initiated impulse) to convulsive-like movements at night, in which the body is tossed from one side of the bed to the other, or repeatedly wheeled around in a circle. Spasms of choking and difficulty in breathing also occur at times during the night. The character of the movements differs from that of the convulsive tics and Sydenham's chorea in being slower and more comprehensive. The motions are easier and wave-like and resemble more closely athetosis, without, however, any tendency toward contraction or fixation of the muscle group at the completion of the movement.

Mental Symptoms.—The ordinary appearance, behavior and conversation are fairly normal. The disposition and temperament, with the exception of the depressed condition at the onset of the disease, has always been happy and contented. Closer examination reveals distinct and decided dementia which is manifested by loss and inaccuracy of memory, both for near and remote events; a child-like simplicity, interest and pleasure in little matters; a lack of appreciation of her condition both as regards the choreic movements and mental deficiencies; a failure of the power to reason or be taught the simplest things, mental and physical. Volition is diminished, but there is an impatient and impulsive desire to follow out any idea, reasonable or otherwise, which comes in her mind. She has no hallucinations or delusions. Her mental condition, in brief, is like that of a child. She is very happy to the point of a mildly exalted state. Busies herself reading and fussing over little details in her daily routine. Enjoys social life, but is equally content to be alone.

Treatment.—Arsenic, bromide, hyoscine, hyosciamus and silver nitrate in ordinary doses had no effect on the chorea. In larger, paralyzing doses some reduction of the movements was noticed, but the reaction after ceasing their use was worse than before. Rest, isolation and the hygienic regulation of the daily life gave the best result, producing a temporary improvement noticeable both in the physical and mental conditions. After the experimentation with drugs was over and she went on the rest treatment the patient gained twenty-three pounds.

The following psycho-physiological considerations based upon the symptoms presented by this patient will be of service in correctly interpreting the true nature of Huntington's chorea.

Conceiving the arc of sentient life, or physical manifestations, to consist of three parts, we have on one side the flow of afferent stimulations from the periphery to the coördinating centres of the central nervous system, and on the other side, the sending forth from these centres efferent impulses resulting in motor activity. The nerve centres, spinal and encephalic, occupy the middle point of the arc. Examination of the nerves of the special senses and the general sensibility shows that the peripheral stimuli are, in the main, normally received and transmitted. Likewise testing the motor apparatus reveals the fact that the motor nerves transmit the efferent impulses, such as they are, properly and that the muscles are in good order and respond correctly to these impulses.

The cause for the irregular and abnormal behavior of the muscles, therefore, does not lie in the muscles themselves nor in their innervation, but in the peculiar character of the impulses they receive. These impulses arise in the nerve centres, hence we may conclude that here is the true seat of the disorder. The nerve centres chiefly involved are those of the cortical and subcortical motor areas and, disregarding the variable and coarser anatomical findings, the essential lesion, as described by Oppen-

heim¹, Dana², Michell Clarke³, Facklam⁴ and Collins⁵, is of the nature of a chronic, parenchymatous encephalitis.

The symptoms of this case support this view of the pathology of the disease in two respects, one is the fact that direct psychical influences are the most potent causes of the choreic manifestations; any emotional excitement or stimulation of the mental faculties greatly exaggerating the movements. The other is the complete change of consciousness, or ego of the individual, and the signs of distinct mental deterioration manifested in the form of a progressive dementia.

With comparatively few exceptions the histories of reported cases state that the chorea was noticed first and the mental symptoms appeared later. It is natural that this should be so, because the chorea as a rule is the conspicuous and absorbing symptom which brings the patient to the notice of the physician. Added to this is the fact that the disorder is quite commonly thought of and treated as a neurosis rather than psychosis, and the mental aspect of the case is consequently not apt to be so thoroughly investigated. (Charcot⁶, Lannois⁷, Jolly⁸.)

The writer is of the opinion that if more care were taken in studying the life history of the patient it would be found in a large number of cases that at the time of onset, if not before, distinct changes in the mental condition were already present. Huber⁹ was one of the first to report a case showing clearly the signs of mental deterioration previous to the appearance of the chorea. Several other cases have been similarly reported and re-

¹ Arch. f. Psych., xxv., 3.

² Journal Nerv. and Ment. Dis., Sept., 1895.

³ Brain, xx., 77, 1897.

⁴ Arch. f. Psych., xxx., 1, 1897.

⁵ Boston Med. and Surg. Journal, Dec. 23, 1897.

⁶ Lecons du Mardi. Policlinique, I. and II., 1887 and 1888.

⁷ Rev. de méd., Aug., 1888. Le bulletin méd., Nov., 1894.

⁸ Neurol. Centralblatt, x., June, 1891.

⁹ Virchow's Archiv., cviii., 1 and 2.

cently Facklam¹⁰, in an extended review of the disease, states that five of his eight cases showed previous mental change.

A comparative study of the different kinds of motor disturbance in connection with the various lesions of the central nervous system would be of distinct value in broadening our conception of such a disease as Huntington's chorea. Such a study should induce, on the one hand, the neurologist to consider cases of tic, Friedreich's ataxia, paralysis agitans, athetosis, etc., more from the psychological standpoint than is at present done, and on the other hand, the alienist would be induced to attach more significance to the play of the motor symptoms in the insane.

To illustrate the importance of taking a comprehensive view of the disorders of motility I will cite a case of generalized tic, characterized by pronounced motor symptoms with, as commonly considered, almost no mental peculiarities. In support of the view that the convulsive tics are, in the main, a degenerative disorder, we will find that the majority of cases, even of mild type, show certain psychical defects or departures in mental action from the normal standard. Noting, for instance, that the body and limb movements are rapid and changeable we will discover that the mental functions are similarly affected. The patient is apt to be restless and impulsive in conduct, finding it difficult to keep the attention concentrated on one line of intellectual work. The memory is often capricious and there frequently exists a superficial conception of the affairs of life. The mind is hurried and irregular in action just as the muscular movements. In fact the movements are almost exactly paralleled by the mental habits. Pursuing the examination further will reveal other associate imperfections of the psychical state.

My purpose in alluding in this connection to such a

¹⁰ Loc. cit.

disease as convulsive tic is to bring out the fact, first, that motor activity is the outward expression of the state of the motor centres, and secondly, that these centres are intimately connected by association fibres with those of higher consciousness¹¹ and, therefore, there is good reason to believe that whenever we find distinct and permanent motor disturbance of cerebral origin we may also expect to meet a certain kind or grade of mental deficiency or impairment. This impairment will vary according to the degree and type of the affection. It is true that it need not necessarily be present at all, as it is possible to conceive that the motor centres alone functionate imperfectly without involvement of the higher centre association fibres. The rule is, however, as Arndt¹² has clearly stated, that we will generally find some mental deficiency if we study carefully the habits, characteristics and inner life of the individual. Ordinary clinic or private visits, unless very frequent, often fail to reveal the facts of this kind. One must almost live with his patient to fully appreciate the psychical side of many of the so-called neuroses. Hence, in spite of the criticism, and citation of a case to the contrary, of so thorough a student of chorea as Sinkler¹³, we are forced by psychological reasons to believe that Diller¹⁴ is right in asserting that chorea long continued is almost invariably connected with mental abnormalities.

In the case of tic we may suppose the motor centres to be chiefly affected with slight involvement of the adjoining or association centres. In ordinary chronic dementia these adjoining centres of ideation, etc., are deeply affected, while the motor centres themselves are comparatively perfect in function, except as they are brought in

¹¹ Flechsig (L. F. Barker): "Sense Areas and Association Centres." *Journal Nerv. and Ment. Dis.*, xxiv., 6.

¹² *Arch. f. Psych.*, I., 3, p. 509.

¹³ *N. Y. Med. Record*, March, 1892.

¹⁴ *Am. Journal Med. Sc.*, April, 1890.

play secondarily, as is frequently seen in the rhythmic movements or habitual body posturing of such patients. Tic, paralysis agitans and athetosis manifestly cannot be considered psychoses, if we abide by the definition of the term, but Huntington's chorea, if we place it in the category with dementia paralytica and dementia senilis, and view it as an affection of the combined motor and association centres, may unquestionably be classed as a psychosis.

In determining whether the term "dementia" is applicable to Huntington's chorea it will be noticed that the point of view makes a difference in our conception. The neurologist chiefly interested in the motor symptoms is apt to underrate the picture of psychical decay before him, while with the student of insanity, if he has given the subject thought, there is far less difficulty in appreciating the unmistakable features of a true dementia with the disorder of motility occupying a position of secondary importance. Phelps¹⁵ makes an excellent plea in this latter direction, and a further witness on the side of the alienist's conception is the reviewer of Osler's "Treatise on Chorea" in the *Journal of Mental Science*, April, 1895, in which he raises the question: Why not a general chorea, as well as a general paralysis of the insane? It is interesting to note in this connection that Golgi¹⁶ pointed out, in 1874, the similarity in pathological lesions between chronic hereditary chorea and progressive paralysis. Since then this comparison has been made by several others. (Hoffmann¹⁷, Phelps¹⁸, Bondurant¹⁹.)

Setting aside for the moment the question of the propriety of the designation, dementia, we need be in no doubt as to the prognosis, course, futility of treatment and termination of Huntington's chorea. Given the

¹⁵ *Journal Nerv. and Ment. Dis.*, Oct., 1892.

¹⁶ *Rivista Clinica*, 1874 (abs. *Jo. Ment. Sc.*, xxii., p. 322).

¹⁷ *Virchow's Archiv.*, cxi., 3, p. 513.

¹⁸ *Hosp. Bull.*, Second Minnesota, 1892.

¹⁹ *Journal Nerv. and Ment. Dis.*, Oct., 1896.

heredity and the characteristic chorea appearing in middle life, the prognosis can be stated with a surety fully equal to that of dementia paralytica and dementia senilis. The slowness of onset of the mental deterioration and the cases exhibiting melancholic or maniacal symptoms do not in the least preclude the prognosis, and so in reality a diagnosis of progressive dementia.

One of the chief difficulties in considering hereditary chorea a form of dementia lies in the variation in type which is noticed to exist among reported cases. Thus, the cases cited by Hoffmann²⁰ and by Chaufford²¹, of chronic hereditary chorea without psychic disturbance make one question the completeness of their observations; the patient of Sinkler²², 34 years old, choreic from birth, and normal mentally; the two cases of Bower²³, one 36 years old, choreic at 30, with previous history of alcoholism and paraplegic attack, the other, 32 years old, with chorea of three years standing, not hereditary, and no mental symptoms; these and such other manifestations of chronic progressive chorea as have been observed must be considered as exceptions to the general type under consideration. The special character of the degenerative process and its manner of attacking the neuron, the rate of its progress and the locality of the tracts and areas affected must explain in this, as in all similar diseases, such variations as the above. Until more recently the cases have not been seen, or for some other reason, European authors seem to have had more difficulty in recognizing such a distinct form of hereditary chorea as exists in this country. Certainly American writers have portrayed a type, such as this case represents, with singular uniformity, and it is

²⁰ Loc. cit.—Questioned by Herringham, *Brain*, xi., p. 416; also by Sinkler, *Journal Nerv. and Ment. Dis.*, xiv., p. 80.

²¹ *Le bulletin méd.*, April, 1896.

²² *Med. Record*, March, 1892.

²³ *Journal Nerv. and Ment. Dis.*, March, 1890.

only to cases resembling this class that the term dementia is strictly applicable.

It may be argued that the term dementia can be applied with equal right to epileptic as well as hereditary choreic conditions. Practically this is just what is done when the dementia is well-defined, but the term is not serviceable earlier in the disease, as it is in the case of Huntington's chorea, because the appearance, course and duration of the mental deterioration cannot be foretold with satisfactory definiteness. The variable frequency of the epileptic attacks prevents the accuracy of prognosis, such as is attainable when we are dealing with a non-intermittent, steadily progressive process of degeneration such as exists in general paralysis, dementia senilis and Huntington's chorea.

This cursory review of the extent to which the body movements may express the state of the motor and associated higher consciousness centres must arouse interest in the effect of inherited tendency to degeneration as an etiological factor in the disorders of motility. Thus, it will be noted that in such diseases as tic, paralysis agitans, athetosis, Friedreich's ataxia, Huntington's chorea and dementia senilis the decadence of the neuron is more slowly progressive and is marked by a disturbance rather than loss of the power of movement. The paralysis, if present, is relatively less prominent and does not become pronounced until later. In dementia paralytica and other diseases of the brain and cord which are secondary infections, or to some other positive intercurrent process, the progress of the affection is more rapid and there is a combination of both disorder and paralysis of motion, the latter appearing earlier and as a more distinct symptom than is the case when the neuron loses its normal functions as the result of a natural mal-development or premature degeneration.

In conclusion it may be stated that a comprehensive

view of Huntington's chorea of the type here reported based upon the clinical, pathological and psychological data at our command warrants the following assertions:—first, that it is a progressive, degenerative disease of the brain, fundamentally different from ordinary or Sydenham's chorea; secondly, that the chief physical manifestations of the disease are the choreiform movements which are of secondary importance, merely indicating the nature of the cerebral lesion; thirdly, that the character of the mental symptoms can best be described by the term dementia; and fourthly, if the above assertions can be verified, then the disorder should be classed with such diseases as dementia paralytica and dementia senilis.

It may seem a stretch of the imagination to compare the tremor and other motor disorders of senility to the choreiform movements, but if we conceive of a degenerative process thrust violently upon the individual in middle life, the resemblance between the two diseases is not so unlike. Pathologically this is certainly what occurs, and the view here maintained is well set forth by Dana²⁴ in speaking of the disease as a teratological defect.

The propriety, as well as the advisability, of applying the name "dementia choreica" to Huntington's chorea is of course open to question, but if the term proposed excites a broader conception of the disease than is at present taken and directs attention to the psychological significance of the disorders of motility it will have served its purpose.

DISCUSSION.

Dr. J. J. Putnam thought that the view presented was a broadening one to the mind. Whether or not we had to do in such processes with joint conditions was always open to doubt. One saw the occurrence of various physical stigmata side by side with physiological and psychological evidence of degeneration, and sometimes without them. The same was true with regard to these morbid processes, but when the collocation

²⁴ Journal Nerv. and Ment. Dis., Sept., 1895.

became a fairly definite one, it was highly important to recognize it. What appealed to him most, perhaps, was the statement regarding a logical connection between disorders of movement and the psychical defects. Possibly that gap was not so wide as one might suppose.

Dr. Sinkler said, with regard to the renaming of the disease, that he had always thought the name "Huntington's chorea" was undesirable, both on account of the fact that, in general, the naming of diseases after individuals is undesirable, and because Huntington was not the first writer to describe this peculiar form of hereditary chronic chorea. It had been described as long ago as 1841 in Dunglison's "Practice of Medicine" by Waters, and in 1863 by Lyon. The name which Dr. Hallock proposed was, perhaps, also undesirable, because it led to a risk of confusion with chorea insaniens. He thought also that the writer made too sweeping an assertion when he said that all chorea of long standing leads to mental disorder. He quite agreed with him, however, that in the hereditary type, mental symptoms sooner or later develop without fail, but it is the rule that the mental symptoms develop long after the choreic symptoms begin.

Dr. Joseph Collins said it seemed to him that there was a more important reason than that given by the last speaker why Huntington's chorea should not be rechristened, according to the suggestion in the paper, namely, that in ordinary Sydenham's chorea there is often slight dementia. In fact, in every case of chorea of any considerable intensity and duration there is always a slight grade of dementia, described in the books as hebetude, mental sluggishness and the like, but which is after all a slight dissociation of the components of the mind. Hence, he could not see the propriety in singling out Huntington's chorea and calling it dementia choreica. There might be some who did not call up the clinical picture of mental and motor disturbance when the eponymic nomenclature was employed, but they were certainly very few. If we are bound to change the name of this disease he preferred to wait until we can designate by a term that shall encompass the pathological progress, and, perhaps, also the distinctive clinical features.

With regard to the basic lesion of Huntington's chorea, he had so recently written upon the subject that he would only say that it seemed to him fairly well proven, considering the findings in his own case, and in those of Dana and Oppenheim, which were all parallel, that the disease is a form of chronic parenchymatous encephalitis.

Dr. C. K. Mills thought the paper of Dr. Hallock was a

suggestive one, and, on the whole, the name proposed was a good one. The objections which had been offered to it by the previous speakers would hardly hold on a careful study of the matter. Chorea insaniens is a term applied to a definite form of acute or subacute mental disorder, which is distinctly different from that found in Huntington's chorea, and while some mental disorder is frequently present in many cases of chorea, using the word dementia in the technical sense, he scarcely thought it could be said that even prolonged cases of Sydenham's chorea lead to true dementia. Other forms of disease besides Huntington's chorea may do this. The paper is another evidence of the increasing interest being taken in the integrating of the numerous facts which we now have regarding the nervous system. With regard to the diseases that Dr. Hallock would associate with Huntington's chorea, it seemed to him unwise to class the latter with dementia paralytica. In dementia paralytica there were certain pathological conditions which in most cases were due to acquired disease of a peculiar type, the indirect cause frequently being syphilis.

Dr. Hallock, in closing the discussion, said that he thought Dr. Mills had answered the chief objections raised by Drs. Sinkler and Collins against the use of the name. In his own mind, viewing the question of psychical degeneration from the teratological standpoint, he was inclined to divide the process into three general grades: First, into the degeneration occurring in early life, represented by the adolescent insanities and mental abnormalities; second, into the degeneration in middle life, such as Huntington's chorea; third, into that which occurs at the climacteric period or later in life, represented by dementia senilis. Certainly the type of dementia senilis, which is more purely psychic and not markedly arteriosclerotic, can be considered, in one sense at least, a teratological defect, only further removed in lifetime than is the case with Huntington's chorea.

252. ZUR FRAGE VON DEN LAEHMUNGERSCHINUNGEN BEI PASTEUR'SCHEN IMPFUNGEN (Paralytic Symptoms following Pasteur Injections). L. O. Darkschewitsch (Neurologisches Centralblatt, 17, 1898).

The author presents the histories with clinical notes of two cases of paralysis following Pasteur's treatment of hydrophobia. The main symptoms were those of paresis of the extremities with paræsthesia pain and ataxia. The electrical reactions were less pronounced and there was some atrophy of the small muscles of the hands in one of the cases. In the second case there was developed a double facial paralysis.

Both cases recovered, though the first case retained some paræsthetic symptoms with slight ataxia of the fingers of the left hand for over a year.

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